

IN THE UNITED STATES PATENT AND TRADEMARK OFFICE



Applicants: Thomas J. Hudson, James Engert and Andrea Richter  
Application No.: 09/693,205 Group: 1646  
Filed: October 20, 2000 Examiner: Not yet assigned  
For: Identification of ARSACS Mutations and Methods of Use Therefor

CERTIFICATE OF MAILING	
I hereby certify that this correspondence is being deposited with the United States Postal Service with sufficient postage as First Class Mail in an envelope addressed to Assistant Commissioner for Patents, Washington, D.C. 20231	
on <u>4/5/01</u>	<u>Paula Depelteau</u>
Date	Signature
<u>PAULA DEPELTEAU</u>	
Typed or printed name of person signing certificate	

REPLY TO NOTICE TO FILE MISSING PARTS OF APPLICATION

Box Missing Parts  
Assistant Commissioner for Patents  
Washington, D.C. 20231

Sir:

In reply to the Notice to File Missing Parts dated December 14, 2000, the following documents and fees are being submitted for filing in the captioned application. A copy of the Notice is attached.

☒ **EXECUTED DECLARATION/POWER OF ATTORNEY**

*(Separate transmittal letter and postcard not required)*

☒ **FILING FEE** - with Fee Transmittal for Patent Applications in duplicate.

*(Separate transmittal letter and postcard not required)*

☒ **SURCHARGE** - surcharge fee of \$130.00.

*(Separate transmittal letter and postcard not required)*

☐ **SEQUENCE LISTING** - Filed concurrently and is attached.

*(Separate transmittal letter and postcard required)*

☐ **TRANSMITTAL OF SUBSTITUTE DRAWINGS**

☐ Sheets of substitute drawings consisting of Figs. 1 - ☐ are enclosed.  
(Separate transmittal letter and postcard not required)

☒ **PETITION FOR EXTENSION OF TIME**

☒ Applicants hereby petition to extend the time to respond to the Notice to File Missing Parts dated December 14, 2000 for two months from February 14, 2001 to April 14, 2001. The appropriate fee of \$390 is included in the enclosed check.

☐ A ☐ month extension of time to respond to the Notice to File Missing Parts dated ☐ was filed on ☐ with payment of a \$☐ fee.

☐ Applicant hereby petitions for an additional ☐ month extension of time to respond to the Notice to File Missing Parts. The appropriate fee of \$☐ is included in the enclosed check.

(Separate Petition for Extension of Time and postcard not required)

☒ **REQUEST FOR CORRECTED FILING RECEIPT** - Filed concurrently and is attached.

(Separate transmittal letter and postcard required)

☐ **STATEMENT CLAIMING SMALL ENTITY STATUS**

☐ Was filed on ☐.

☐ Is enclosed herewith.

(Separate transmittal letter and postcard not required)

☐ In view of the small entity status of the captioned application, we hereby request a reimbursement of 50% of the filing fees in the amount of \$☐ which were paid on ☐ to be deposited in Deposit Account No. 08-0380.

The fees required for filing the indicated documents are enclosed in the form of a check in the total amount of \$2,612.00. Please charge any deficiency or credit any overpayment in the fees that may be due in this matter to Deposit Account No. 08-0380. A copy of this letter is enclosed for accounting purposes.

Respectfully submitted,

HAMILTON, BROOK, SMITH & REYNOLDS, P.C.

By

Lisa M. Treannie  
Lisa M. Treannie

Registration No.: 41,368

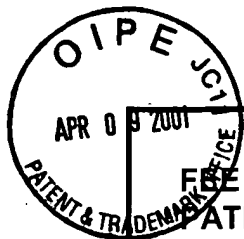
Tel.: (781) 861-6240

Fax: (781) 861-9540

Lexington, Massachusetts 02421-4799

Date:

4/5/01



## HAMILTON, BROOK, SMITH &amp; RENOIDS, P.C.

# 4

FREE TRANSMITTAL FOR  
PATENT APPLICATIONS

Attorney Docket Number	2825.1021-003
Application Number	09/693,205
First Named Inventor	Thomas J. Hudson

CLAIM CALCULATION (includes any preliminary amendment)

CLAIMS	(1) FOR	(2) NUMBER FILED	(3) NUMBER EXTRA	(4) RATE	(5) CALCULATIONS
	TOTAL CLAIMS (37 CFR 1.16(c) or (j))	39 - 20* =	19	x \$ 18 =	\$ 342
	INDEPENDENT CLAIMS (37 CFR 1.16(b) or (i))	16 - 3** =	13	x \$ 80 =	\$ 1040
	MULTIPLE DEPENDENT CLAIMS (if applicable) (37 CFR 1.16(d))			+ \$ 270 =	\$
				BASIC FEE (37 CFR 1.16(a) or (h))	\$ 710
				Total of above Calculations =	\$ 2092
	Reduction by 50% for filing by small entity (37 CFR 1.9, 1.27, 1.28) =				\$
	TOTAL =				\$ 2092
	Surcharge - Late Filing of Declaration or Filing Fees (37 C.F.R. 1.16(e)) =				\$ 130
	Petition for Extension of Time Fee (37 C.F.R. 1.17) =				\$ 390
	Assignment Recordation Fee = (only when filed with application)				\$
	* Reissue claims in excess of 20 and over original patent ** Reissue independent claims over original patent			TOTAL =	\$ 2612

1. Small entity status:

04/11/2001 JADD01 00000024 09693205

05 FC:116

390.00 0P

a. ☐ A small entity statement is enclosed.b. ☐ A small entity statement was filed in the prior non-provisional application and such status is still proper and desired.c. ☐ Is no longer claimed.2. ☒ A general authorization is hereby granted to charge deposit account number 08-0380 for any fees required under 37 CFR 1.16 and 1.17 in order to maintain pendency of this application. A copy of this authorization is enclosed for accounting purposes.3. ☒ A check is enclosed for \$2612.00. ☐ Please charge \$[ ] to Deposit Account No. 08-0380.4. ☐ Other: \_\_\_\_\_

Signature

Lisa M. Treannie

Date

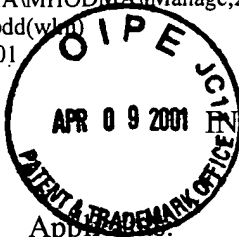
April 5, 2001

Submitted by  
Typed or Printed Name

Lisa M. Treannie

Reg. Number

41,368



IN THE UNITED STATES PATENT AND TRADEMARK OFFICE

Applicant: Thomas J. Hudson, James Engert and Andrea Richter  
Application No.: 09/693,205 Group: 1646  
Filed: October 20, 2000 Examiner: Not assigned  
For: Identification of ARSACS Mutations and Methods of Use Therefor

CERTIFICATE OF MAILING	
I hereby certify that this correspondence is being deposited with the United States Postal Service with sufficient postage as First Class Mail in an envelope addressed to Assistant Commissioner for Patents, Washington, D.C. 20231	
on <u>4/5/01</u>	<u>Paula Depelteau</u>
Date	Signature
<u>PAULA DEPELTEAU</u>	
Typed or printed name of person signing certificate	

REQUEST FOR CORRECTED FILING RECEIPT  
FOR UTILITY APPLICATION

Office of Initial Patent Examination  
Customer Service Center  
Assistant Commissioner for Patents  
Washington, D.C. 20231

Sir:

We hereby request that the following items of information be corrected in the Filing Receipt for the subject application received in this office on December 22, 2000.

The errors and corrections appear below.

The Atty Docket No. for this case should read "2825.1021-003", not "2852.1021-003".

Enclosed are a copy of the Filing Receipt with changes noted in red, a copy of the Utility Patent Application Transmittal, and a copy of page 1 of the Application as filed on October 20, 2000.

Pursuant to instructions in the February 29, 2000 O.G., we hereby request that the errors which are identified above be corrected in the captioned application to which this request for correction is directed. It is understood that the Patent Office will issue an automatically-generated, corrected Filing Receipt in this and, if applicable, any other affected applications.

Respectfully submitted,

HAMILTON, BROOK, SMITH & REYNOLDS, P.C.

By Lisa M. Treannie

Lisa M. Treannie

Registration No.: 41,368

Telephone: (781) 861-6240

Facsimile: (781) 861-9540

Lexington, MA 02421-4799

Date: 4/5/01



## UNITED STATES PATENT AND TRADEMARK OFFICE

COMMISSIONER FOR PATENTS  
UNITED STATES PATENT AND TRADEMARK OFFICE  
WASHINGTON, D.C. 20231  
www.uspto.gov

APPLICATION NUMBER	FILING DATE	GRP ART UNIT	FIL FEE REC'D	ATTY. DOCKET NO	DRAWINGS	TOT CLAIMS	IND CLAIMS
09/693,205	10/20/2000	1646	0	<del>2862.1021</del> 003	22	39	15

2825.1021-003

## FILING RECEIPT



\*OC000000005621416\*

Doreen M Hogle Esq  
HAMILTON BROOK SMITH & REYNOLDS P C  
Two Militia Drive  
Lexington, MA 02421-4799

Date Mailed: 12/14/2000

Receipt is acknowledged of this nonprovisional Patent Application. It will be considered in its order and you will be notified as to the results of the examination. Be sure to provide the U.S. APPLICATION NUMBER, FILING DATE, NAME OF APPLICANT, and TITLE OF INVENTION when inquiring about this application. Fees transmitted by check or draft are subject to collection. Please verify the accuracy of the data presented on this receipt. If an error is noted on this Filing Receipt, please write to the Office of Initial Patent Examination's Customer Service Center. Please provide a copy of this Filing Receipt with the changes noted thereon. If you received a "Notice to File Missing Parts" for this application, please submit any corrections to this Filing Receipt with your reply to the Notice. When the PTO processes the reply to the Notice, the PTO will generate another Filing Receipt incorporating the requested corrections (if appropriate).

## Applicant(s)

Thomas J. Hudson, residence, NOT PROVIDED;  
James Engert, residence, NOT PROVIDED;  
Andrea Richter, residence, NOT PROVIDED;

## Continuing Data as Claimed by Applicant

THIS APPLN CLAIMS BENEFIT OF 60/160,588 10/20/1999

## Foreign Applications

If Required, Foreign Filing License Granted 12/14/2000

## Title

Identification of arsacs mutations and methods of use therefor

## Preliminary Class

530

Data entry by : WARREN, EFREM

Team : OIPE

Date: 12/14/2000

DEC 22 2000

HAMILTON, BROOK,  
SMITH & REYNOLDS, P.C.

**LICENSE FOR FOREIGN FILING UNDER**  
**Title 35, United States Code, Section 184**  
**Title 37, Code of Federal Regulations, 5.11 & 5.15**

**GRANTED**

The applicant has been granted a license under 35 U.S.C. 184, if the phrase "IF REQUIRED, FOREIGN FILING LICENSE GRANTED" followed by a date appears on this form. Such licenses are issued in all applications where the conditions for issuance of a license have been met, regardless of whether or not a license may be required as set forth in 37 CFR 5.15. The scope and limitations of this license are set forth in 37 CFR 5.15(a) unless an earlier license has been issued under 37 CFR 5.15(b). The license is subject to revocation upon written notification. The date indicated is the effective date of the license, unless an earlier license of similar scope has been granted under 37 CFR 5.13 or 5.14.

This license is to be retained by the licensee and may be used at any time on or after the effective date thereof unless it is revoked. This license is automatically transferred to any related applications(s) filed under 36 CFR 1.53(d). This license is not retroactive.

The grant of a license does not in any way lessen the responsibility of a licensee for the security of the subject matter as imposed by any Government contract or the provisions of existing laws relating to espionage and the national security or the export of technical data. Licensees should apprise themselves of current regulations especially with respect to certain countries, of other agencies, particularly the Office of Defense Trade Controls, Department of State (with respect to Arms, Munitions and Implements of War (22 CFR 121-128)); the Office of Export Administration, Department of Commerce (15 CFR 370.10 (j)); the Office of Foreign Assets Control, Department of Treasury (31 CFR Parts 500+) and the Department of Energy.

**NOT GRANTED**

No license under 35 U.S.C. 184 has been granted at this time, if the phrase "IF REQUIRED, FOREIGN FILING LICENSE GRANTED" DOES NOT appear on this form. Applicant may still petition for a license under 37 CFR 5.12, if a license is desired before the expiration of 6 months from the filing date of the application. If 6 months has lapsed from the filing date of this application and the licensee has not received any indication of a secrecy order under 35 U.S.C. 181, the licensee may foreign file the application pursuant to 37 CFR 5.15(b).

**PLEASE NOTE the following information about the Filing Receipt:**

- The articles such as "a," "an" and "the" are not included as the first words in the title of an application. They are considered to be unnecessary to the understanding of the title.
- The words "new," "improved," "improvements in" or "relating to" are not included as first words in the title of an application because a patent application, by nature, is a new idea or improvement.
- The title may be truncated if it consists of more than 600 characters (letters and spaces combined).
- The docket number allows a maximum of 25 characters.
- If your application was submitted under 37 CFR 1.10, your filing date should be the "date in" found on the Express Mail label. If there is a discrepancy, you should submit a request for a corrected Filing Receipt along with a copy of the Express Mail label showing the "date in."
- The title is recorded in sentence case.

Any corrections that may need to be done to your Filing Receipt should be directed to:

Assistant Commissioner for Patents  
 Office of Initial Patent Examination  
 Customer Service Center  
 Washington, DC 20231





**COPY**

**FILE COPY**

-1-



Date: 10/20/00 Express Mail Label No. EL5525 72736 US

Inventors: Thomas J. Hudson, James Engert and Andrea Richter  
Attorney's Docket No.: 2825.1021-003

## IDENTIFICATION OF ARSACS MUTATIONS AND METHODS OF USE THEREFOR

### RELATED APPLICATIONS

This application claims the benefit of U.S. Provisional application Serial No.  
5 60/160,588, filed October 20, 1999, the entire teachings of which are incorporated  
herein by reference.

### BACKGROUND OF THE INVENTION

Autosomal recessive spastic ataxia of Charlevoix-Saguenay (ARSACS) is an  
early-onset neurodegenerative disease with high prevalence in the Charlevoix-  
10 Saguenay-Lac-Saint-Jean (CSLSJ) region of Quebec. Disease progression is rapid  
through young adulthood, with most patients requiring wheelchairs by their early  
forties. The disease is characterized by abolished sensory nerve conduction, reduced  
motor nerve velocity, and a unique clinical feature of hypermyelination of retinal nerve  
fibers. Additional pathological features include atrophy of the upper cerebellar vermis,  
15 absence of Purkinje cells, and possibly abnormal neuronal lipid storage (Bouchard, J-P.,  
*In: Handbook of Clinical Neurology 16: Hereditary neuropathies and spinocerebellar  
degenerations, J.M.B.V. de Jong, Ed., pp. 451-459, Elsevier Science Publishers,  
Amsterdam (1991)). A developmental defect in the myelination of both retinal and  
peripheral nerve fibers has been proposed as the physiological basis of the disease  
20 (Bouchard, J-P., et al., Neuromuscular Disorders 8:474-479 (1998)). More than 300*



# UTILITY PATENT APPLICATION TRANSMITTAL

(Only for new nonprovisional applications under  
37 C.F.R. 1.53(b))

Attorney Docket No:

2825.1021-503

First Named Inventor or  
Application Identifier

Thomas J. Hudson

Express Mail Label No.

EL552572736US

**COPY****FILE COPY**Title of  
Invention

IDENTIFICATION OF ARSACS MUTATIONS AND METHODS OF USE THEREFOR

**APPLICATION ELEMENTS**

See MPEP chapter 600 concerning utility patent application contents.

ADDRESS TO:

Assistant Commissioner for Patents  
Box Patent Application  
Washington, D.C. 20231

1. ☐ Fee Transmittal Form  
(Submit an original, and a duplicate for fee processing)
2. ☒ Specification [Total Pages 1-57] ☐  
(preferred arrangement set forth below)
  - Descriptive title of the invention
  - Cross References to Related Applications
  - Statement Regarding Fed sponsored R & D
  - Reference to microfiche Appendix
  - Background of the Invention
  - Summary of the Invention
  - Brief Description of the Drawings
  - Detailed Description
  - Claim(s)
  - Abstract of the Disclosure
3. ☒ Drawing(s) (35 U.S.C. 113) [Total Sheets 1-22] ☐  
[ ☒ ] Formal [ ☐ ] Informal
4. ☐ Oath or Declaration/POA [Total Pages [ ] ]
  - a. ☐ Newly executed (original or copy)
  - b. ☐ Copy from a prior application (37 C.F.R. 1.63(d))  
(for continuation/divisional with Box 17 completed)  
[NOTE Box 5 below]
    - i. ☐ **DELETION OF INVENTOR(S)**  
Signed statement attached deleting  
inventor(s) named in the prior  
application, see 37 C.F.R. 1.63(d)(2)  
and 1.33(b).
5. ☐ Incorporation By Reference (useable if Box 4b is checked)  
The entire disclosure of the prior application, from which a  
copy of the oath or declaration is supplied under Box 4b, is  
considered as being part of the disclosure of the accompanying  
application and is hereby incorporated by reference therein.

6. ☐ Microfiche Computer Program (Appendix)
7. ☐ Nucleotide and/or Amino Acid Sequence Submission  
(if applicable, all necessary)
  - a. ☐ Computer Readable Copy
  - b. ☐ Paper Copy (identical to computer copy)  
[ ] Pages
  - c. ☐ Statement verifying identity of above copies

**ACCOMPANYING APPLICATION PARTS**

8. ☐ Assignment Papers (cover sheet & documents)
9. ☐ 37 C.F.R. 3.73(b) Statement [ ☐ ] Power of Attorney  
(when there is an assignee)
10. ☐ English Translation Document (if applicable)
11. ☐ Information Disclosure Statement (IDS)/PTO-1449 [ ☐ ] Copies of IDS  
Citations
12. ☐ Preliminary Amendment
13. ☒ Return Receipt Postcard (MPEP 503)  
(Should be specifically itemized)
14. ☐ Small Entity Statement(s) [ ☐ ] Statement filed in prior application,  
status still proper and desired
15. ☐ Certified Copy of Priority Document(s)  
(if foreign priority is claimed)
16. ☐ Other: \_\_\_\_\_

17. If a **CONTINUING APPLICATION**, check appropriate box and supply the requisite information:[ ☐ ] Continuation [ ☐ ] Divisional [ ☐ ] Continuation-in-part (CIP) of prior application No.:

Prior application information:

Examiner:

Group Art Unit:

**18. CORRESPONDENCE ADDRESS**

NAME	Doreen M. Hogle, Esq.				
	HAMILTON, BROOK, SMITH & REYNOLDS, P.C.				
ADDRESS	Two Militia Drive				
CITY	Lexington	STATE	MA	ZIP CODE	02421-4799
COUNTRY	USA	TELEPHONE	(781) 861-6240	FAX	(781) 861-9540

Signature	<i>Lisa M. Treannie</i>	Date	October 20, 2000
Submitted by Typed or Printed Name	Lisa M. Treannie	Reg. Number	41,368

EL552572736US

#3



UNITED STATES PATENT AND TRADEMARK OFFICE

COMMISSIONER FOR PATENTS  
UNITED STATES PATENT AND TRADEMARK OFFICE  
WASHINGTON, D.C. 20231  
www.uspto.gov

APPLICATION NUMBER	FILING/RECEIPT DATE	FIRST NAMED APPLICANT	ATTORNEY DOCKET NUMBER
09/693,205	10/20/2000	Thomas J. Hudson	2852.1021-003

Doreen M Hogle Esq  
HAMILTON BROOK SMITH & REYNOLDS P C  
Two Militia Drive  
Lexington, MA 02421-4799

**DOCKETED****FORMALITIES LETTER**

\*OC000000005621417\*

Date Mailed: 12/14/2000

**NOTICE TO FILE MISSING PARTS OF NONPROVISIONAL APPLICATION****FILED UNDER 37 CFR 1.53(b)*****Filing Date Granted***

An application number and filing date have been accorded to this application. The item(s) indicated below, however, are missing. Applicant is given TWO MONTHS from the date of this Notice within which to file all required items and pay any fees required below to avoid abandonment. Extensions of time may be obtained by filing a petition accompanied by the extension fee under the provisions of 37 CFR 1.136(a).

- The statutory basic filing fee is missing.  
*Applicant must submit \$ 710 to complete the basic filing fee and/or file a small entity statement claiming such status (37 CFR 1.27).*
- Total additional claim fee(s) for this application is \$1302.
  - \$342 for 19 total claims over 20.
  - \$960 for 12 independent claims over 3.
- The oath or declaration is missing.  
*A properly signed oath or declaration in compliance with 37 CFR 1.63, identifying the application by the above Application Number and Filing Date, is required.*
- To avoid abandonment, a late filing fee or oath or declaration surcharge as set forth in 37 CFR 1.16(e) of \$130 for a non-small entity, must be submitted with the missing items identified in this letter.
- The balance due by applicant is \$ 2142.

*A copy of this notice **MUST** be returned with the reply.*

Customer Service Center

Initial Patent Examination Division (703) 308-1202

PART 2 - COPY TO BE RETURNED WITH RESPONSE

04/11/2001 JADD01 00000024 09693205  
710.00 00 00  
1040.00 00 00  
342.00 00 00  
130.00 00 00  
01 FC:101  
02 FC:102  
03 FC:103  
04 FC:105

12/14/00

